

Transparency and advocacy improve the CF outlook

My daughter was literally starving to death, right in front of our eyes,” recalls Donald Kreis, the father of a young DHMC patient. “Nobody was telling us why that was happening. It was scary for first-time parents, even when one of them is a doctor.” Kreis’s wife is Dr. Jennifer Keller, a 2001 graduate of DMS and now an anesthesiologist at DHMC. Their daughter was only four months old when they noticed that something was wrong with her.

The diagnosis? Cystic fibrosis (CF). Thick mucus blocking the ducts in the baby’s pancreas prevented digestive enzymes from doing their job. “The cure for that is pretty simple,” says Kreis. “You just replace the digestive

enzymes that aren’t getting to the small intestines. We have pills that do that now.”

CF is a chronic, progressive disorder caused by inheriting two copies of a defective gene. Mucous glands in the respiratory, digestive, and reproductive systems produce thick, gluey secretions. People with the disease are prone to chronic lung infections and are unable to absorb fats and other nutrients. In the U.S., about 30,000 children and adults have CF, and more than 10 million people carry one copy of the defective CF gene.

At one time, children with the disease died before they reached first grade. But today—with antibiotics to fight infections, medications to thin mucus, therapies to alleviate symptoms, and better understanding of how to maintain nutrition—people with CF can survive well into adulthood. Kreis’s daughter, now five, takes a number of medications and for 40 minutes a day wears a vibrating vest to loosen the mucus in her lungs.

Data: CF care, as Kreis and Keller have discovered, works best when it’s a partnership between families and caregivers. Dartmouth has been a national leader in spreading the word of that approach. To foster such partnerships, the national CF Foundation recently began publicly reporting health outcomes for more than 115 accredited CF centers. The data ranges from lung function to nutritional status. Dartmouth’s CF center has been publishing such data on

the DHMC website since 2005.

“Dartmouth, I think, has really been at the forefront of seeing the virtues of this kind of transparency,” says Kreis. But, he cautions, “this national transparency is not an occasion for individual CF patients . . . to switch centers if the data from their center troubles them. What it really calls people to do is to . . . figure out how to make your center better able to be as high achieving as the best center.”

Kreis made that point in a recent CF Foundation webcast in which Dartmouth was well represented. Both Kreis and DMS’s Dr. Worth Parker participated in the national discussion about CF care. “The care centers have done remarkable work improving the quality of life for those with the disease,” says DMS’s Dr. Gerald O’Connor, who chaired the expert committee that developed the CF Foundation reporting program. “The reporting of this data is an important sign of the shared responsibility of clinicians and patients.”

Keller and Kreis also served on a national task force to build awareness about the value of family-provider partnerships and were founding members of New Hampshire’s CF Patient/ Family Advisory and Advocacy Council. Among the group’s accomplishments have been lobbying successfully for CF screening of all newborns in the state. Early diagnosis can increase life expectancy. So far about half of the 50 states have newborn CF screening. “I’m working on the other half,” says Kreis.

LAURA STEPHENSON CARTER



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Don Kreis, pictured with his daughter, is combating CF on several fronts.

THEN & NOW

A reminder of the pace of change, and of timeless truths, from 1943’s *Fifty Years of Service*, a history of Mary Hitchcock Memorial Hospital:

“On May 18, 1893, a girl of 13 . . . was admitted to the Mary Hitchcock Memorial Hospital, the doors of which had been opened a few days before. Her difficulty was noted as ‘hip joint disease.’ . . . After a period of observation, an operation was performed, from which she made a slow recovery. On August 8 she was discharged with the notation ‘improved.’ This was case No. 1 in the records of the Hospital. In the 50 years which have since elapsed, some 74,000 sufferers from one physical ailment or another have followed in her train.”



21,254

Number of inpatients discharged from DHMC in 2005 alone